

Contents

- 1 General Description 1**
 - Introduction..... 1
 - Incidence and Prevalence..... 3
 - Predisposing and Genetic Factors 8
 - References..... 13

- 2 Natural History: Importance of Size, Site, and Histopathology 19**
 - Natural History..... 19
 - Influence of Site 19
 - Staging 19
 - Staging of Retroperitoneal Visceral Sarcoma..... 26
 - Prognostic Factors for Extremity and Superficial Soft Tissue Sarcoma..... 30
 - Disease-Specific Survival 31
 - Prognostic Factors for Survival Following Local Recurrence of Extremity Sarcoma 32
 - Prognostic Factors: Nomograms..... 32
 - References..... 34

- 3 General Statement as to Efficacy of Surgery/Chemotherapy/Radiation Therapy 37**
 - Extent of Primary Surgery 37
 - Surgical Treatment of Local Recurrence 39
 - Diagnostic Imaging..... 39
 - Surgery for Metastatic Disease 41
 - Pulmonary Metastasis 41
 - Radiation Therapy..... 49
 - Adjuvant Radiation Therapy..... 49
 - Dose of Radiation Therapy 51
 - Adjuvant Brachytherapy 51

Radiation Therapy in the Presence of Positive Microscopic Margins	52
Definitive Radiation Therapy	52
Radiation to Patients with Neurovascular Involvement.....	52
Wound Complications of Radiation Therapy.....	53
Sarcomas More Common in Adults	54
Larger Randomized Studies	54
Selected Meta-analyses of Randomized Trials of Adjuvant Chemotherapy	56
Adjuvant Therapy for GIST	57
Sarcomas More Common in the Pediatric Setting	58
Special Techniques for Primary and Locally Recurrent Disease	60
Intra-arterial Chemotherapy	60
Limb Perfusion and Hyperthermia.....	60
References.....	61

Part I Management by Histopathology: Introduction

4 Gastrointestinal Stromal Tumors (GISTs).....	67
Imaging	67
Familial GIST	67
Natural History.....	72
Diagnosis, Molecular Pathology	73
Treatment	74
Adjuvant Imatinib for Primary GIST.....	74
Neoadjuvant Therapy for Primary Disease Not Amenable to Surgery	77
Treatment of Recurrence.....	77
Systemic Targeted Therapy for Metastatic GIST.....	77
Dose Intensity over Time	82
Imatinib Pharmacokinetics.....	83
Second-Line Sunitinib for Imatinib-Resistant Metastatic GIST.....	84
Other Tyrosine Kinase Inhibitors for Metastatic GIST Failing Imatinib and Sunitinib	85
Newer Agents.....	85
References.....	87
5 Liposarcoma	93
Imaging	97
Diagnosis.....	99
Treatment	99
Radiation Therapy.....	103
Systemic Therapy: General Considerations	104
Adjuvant Therapy	105
Treatment of Metastatic Disease.....	105

Outcomes	107
Outcome Following Metastasis.....	109
References.....	110
6 Leiomyosarcoma	113
Imaging	113
Diagnosis, Molecular Pathology	115
Treatment	117
Radiation Therapy	120
Systemic Therapy.....	120
Adjuvant Chemotherapy	120
Treatment of Recurrence.....	121
Metastatic Disease	121
Outcome.....	123
References.....	124
7 Undifferentiated Pleomorphic Sarcoma	
(UPS; Malignant Fibrous Histiocytoma: MFH)	
and Myxofibrosarcoma.....	129
Imaging	129
Diagnosis/Molecular Pathology	129
Natural History.....	131
Treatment	132
Radiation Therapy.....	132
Metastatic Disease	132
Adjuvant Chemotherapy	133
Outcome.....	134
References.....	135
8 Synovial Sarcoma.....	137
Imaging	137
Diagnosis, Molecular Pathology	137
Treatment	141
Radiation Therapy.....	141
Chemotherapy	141
Treatment of Recurrence.....	143
Local Recurrence	143
Systemic Treatment.....	143
Outcome.....	145
References.....	146
9 Malignant Peripheral Nerve Sheath Tumor	
(MPNST) and Triton Tumor	149
Presentation.....	149
Imaging	149
Diagnosis/Pathology	152
Neurofibromatosis Type 1 and Outcome	154

Treatment	154
Radiation Therapy	154
Chemotherapy	155
Outcome	157
References	158
10 Desmoid Tumor/Deep-Seated Fibromatosis	
(Desmoid-Type Fibromatosis)	161
Clinical Presentation	163
Imaging	163
Diagnosis: Molecular	163
Natural History	165
Treatment	166
Treatment of Recurrence	166
Systemic Therapy	167
Treatment by Observation	169
Radiation Therapy	169
Patterns of Failure	170
Outcome	171
References	175
11 Solitary Fibrous Tumor/Hemangiopericytoma	179
Outcome	182
References	183
12 Fibrosarcoma and Its Variants	185
Outcome	185
Dermatofibrosarcoma Protuberans	185
Outcome	188
Low-Grade Fibromyxoid Sarcoma (Evans' Tumor)	189
Outcome	194
Sclerosing Epithelioid Fibrosarcoma	194
Inflammatory Myofibroblastic Tumor	195
Infantile Fibrosarcoma	196
Myxoinflammatory Fibroblastic Sarcoma/Inflammatory	
Myxohyaline Tumor of Distal Extremities	197
Adult-Type Fibrosarcoma	197
References	198
13 Vascular Sarcomas	201
Epithelioid Hemangioendothelioma	201
Angiosarcoma/Lymphangiosarcoma	204
Outcome	209
Kaposi Sarcoma	210
References	211

14	Epithelioid Sarcoma	215
	Outcome.....	217
	References.....	219
15	Sarcomas More Common in Children	221
	Soft Tissue Sarcomas More Commonly Observed in Pediatric Patients.....	221
	Ewing Sarcoma Family of Tumors (EFT)	222
	Demographics	226
	Primary Therapy	226
	Adjuvant Chemotherapy	228
	High-Dose Systemic Therapy for Metastatic Disease	229
	Standard Cytotoxic Chemotherapy After Disease Relapse.....	230
	Investigational Approaches	231
	Ewing Sarcoma-Like Small Round Cell Tumors.....	232
	Rhabdomyosarcoma.....	233
	Demographics	234
	Molecular Biology	236
	Risk Stratification.....	238
	Staging	238
	Imaging	240
	Primary Therapy	240
	Chemotherapy for Metastatic Disease	242
	Embryonal Sarcoma.....	243
	Calcifying Nested Stromal and Epithelial Tumor.....	244
	References.....	245
16	Radiation-Induced Sarcomas	251
	References.....	256
17	Alveolar Soft Part Sarcoma	259
	Imaging	259
	Diagnosis/Molecular Pathology	259
	Primary Treatment	262
	Treatment of Metastatic Disease.....	262
	Outcome.....	263
	References.....	264
18	Clear Cell Sarcoma/Melanoma of Soft Parts	267
	Imaging	267
	Diagnosis: Molecular Pathology.....	267
	Treatment	270
	Outcome.....	271
	References.....	272

19	Desmoplastic Small Round Cell Tumor	275
	Imaging	275
	Diagnosis.....	275
	Treatment	276
	Outcome.....	278
	References.....	279
20	Extraskelatal Myxoid Chondrosarcoma	281
	Imaging	281
	Diagnosis.....	281
	Treatment	285
	Outcome.....	285
	References.....	286
21	Other Uterine Sarcomas	289
	Endometrial Stromal Sarcoma	289
	Diagnosis.....	289
	Treatment	290
	Outcome.....	291
	Undifferentiated Endometrial Sarcoma	293
	Outcome.....	294
	Uterine Carcinosarcomas and Other Malignant	
	Mixed Müllerian Tumors	294
	PEComas.....	296
	Outcome.....	296
	References.....	297
22	Extraskelatal Osteogenic Sarcoma	301
	Imaging	301
	Diagnosis.....	301
	Treatment	305
	Outcome.....	305
	References.....	307
23	Sustentacular Tumors of Lymph Tissue.....	309
	Follicular Dendritic Cell Tumor (Dendritic Reticulum	
	Cell Tumor [FDCT]) and Interdigitating Reticulum	
	Cell Tumor (IDRCT).....	312
	True Histiocytic Sarcoma.....	312
	Langerhans Cell Tumors	313
	Outcome.....	313
	References.....	315
24	Uncommon/Unique Sites	317
	Heart and Great Vessels	317
	Primary Sarcomas of the Breast.....	318
	Phyllodes Tumor	321
	Diagnosis.....	322
	Outcome.....	323

Head and Neck	323
Treatment	324
Primary Sarcomas of the Mediastinum.....	324
Liver.....	325
References.....	325

Part II Benign and Less Aggressive Lesions: Introduction

25 Mostly Benign/Rarely Metastasizing Soft Tissue Tumor	329
Ossifying Fibromyxoid Tumor	329
Perivascular Epithelioid Cell Tumor (PEComa) and Related Entities: Lymphangiomyomatosis, Angiomyolipoma, and Sugar Cell Tumor.....	330
Therapy	331
Giant Cell Tumor of Tendon Sheath/Pigmented Villonodular Synovitis	332
Myoepithelioma of Soft Tissue.....	334
Glomus Tumor	334
References.....	336
26 Benign Soft Tissue Tumors.....	339
Lipoma	339
Lipomatosis.....	340
Lipoblastoma/Lipoblastomatosis	340
Angiolipoma	341
Angiomyolipoma	341
Angiomyelolipoma	343
Hibernoma.....	343
Elastofibroma	345
Granular Cell Tumor	347
Hemangioma	347
Leiomyoma	350
Schwannoma	350
Neurofibroma	350
Myxoma	351
Angiomyxoma.....	352
References.....	353
27 Reactive Lesions	355
Myositis Ossificans	355
Nodular Fasciitis	355
Sarcoma Masquerade	356
References.....	358
Suggested Keywords	359
Index.....	369



<http://www.springer.com/978-1-4614-5003-0>

Management of Soft Tissue Sarcoma

Brennan, M.F.; Antonescu, C.R.; Maki, R.G.

2013, XV, 380 p., Hardcover

ISBN: 978-1-4614-5003-0